Thoracic ectopic kidney in a child: a case report

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Congenital thoracic ectopic kidney is a very rare developmental anomaly and the rarest form of all ectopic kidneys. It is usually asymptomatic and discovered incidentally on a routine chest radiography. We report a thoracic ectopic kidney in a 19-month-old boy, which initially presented as a well demarcated mass at the base of the right lung on chest x-ray. Intravenous pyelography (IVP) and thoraco-abdominal computed tomography (CT) demonstrated a normal functioning transdiaphragmatic thoracic ectopic right kidney, but technetium-99m DTPA and DMSA scintigraphy demonstrated pelvic stasis. We hereby discuss the features of congenital thoracic ectopic kidney and review the literature. Although it is extremely rare, thoracic ectopic kidney should be considered in differential diagnosis of a mass with a well demarcated superior margin in the lower part of the thorax, and renal scintigraphy must be performed even if CT and IVP results are normal.

Key words: thoracic ectopic kidney.

Congenital thoracic ectopic kidney is an uncommon developmental anomaly and the rarest form of all ectopic kidneys[1]. Of nearly 16,000 autopsies performed in a series, 22 ectopic kidneys were found, and only one was intrathoracic[2]. Wolfromm[3] reported the first case of clinically diagnosed congenital ectopic kidney by retrograde pyelography in a 43-year-old patient in 1940. It is generally asymptomatic and discovered incidentally on a routine chest radiography[1].

We herein report a thoracic ectopic kidney in a 19-month-old boy, which initially presented as a well demarcated mass at the base of the right lung on chest x-ray.

Case Report

A 19-month-old male infant had initially been examined for cough and wheezing of seven days' duration in a peripheral hospital. At that time, his chest x-ray had revealed a well demarcated homogeneous mass at the base of the right lung (Fig. 1). The patient was refererd to our hospital for evaluation of that mass. Prenatal, natal, postnatal and developmental history was unremarkable. Vital signs including body temperature, heart rate, blood pressure, and respiration rate and pattern were normal. Physical examination was normal except for his serous nasal discharge. Abdominal ultrasonography showed that the right kidney was located superior to its normal location (Fig. 2). Thoraco-abdominal computed tomography demonstrated a transdiaphragmatic thoracic ectopic right kidney (Fig. 3). Intravenous pyelography (IVP) revealed the mass in the right hemithorax to be an ectopic intrathoracic kidney with a normal collecting system and function (Fig. 4). Technetium-99m DTPA and DMSA scintigraphy demonstrated that the right kidney was superior to its normal location and draining spontaneously with pelvic stasis (Fig. 5).

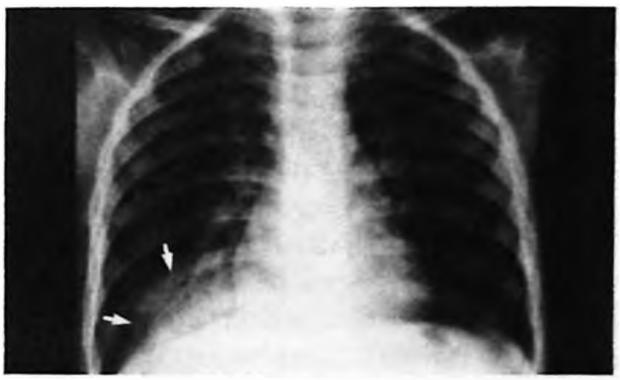

Fig. 1. Chest radiography of patient reveals a well demarcated homogeneous mass in the base of the right hemithorax.

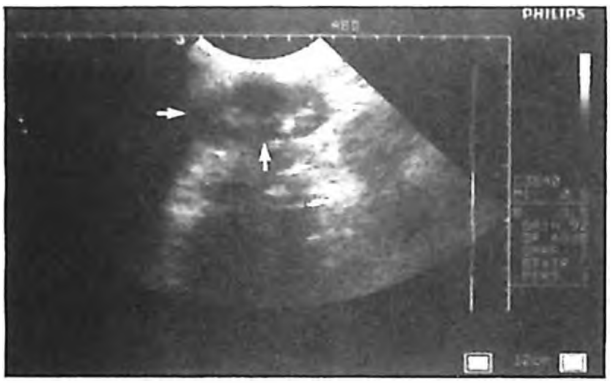

Fig. 2. Ultrasonography of the mass in the right hemithorax through posterior thoracic wall reveals right kidney.

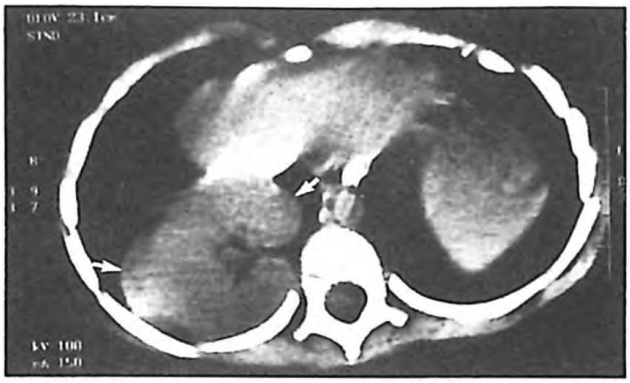

Fig. 3. Thoraco-abdominal computed tomography shows abnormal intrathoracic location of right kidney with remarkable calyceal system.

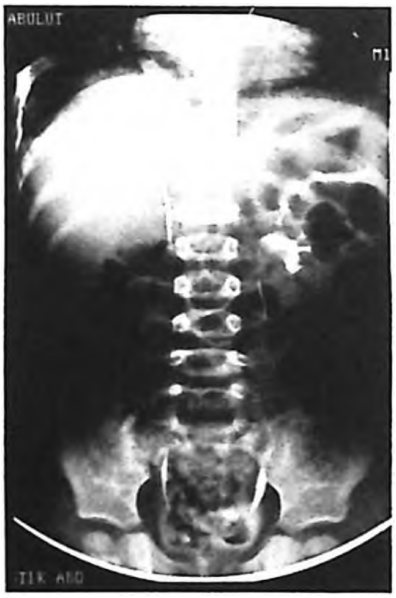

Fig. 4. Intravenous pyelography reveals the mass in the right hemithorax to be an ectopic intrathoracic kidney with a normal calyceal system, ureter and function.

Fig. 5. A postero-anterior view of technetium-99m DTPA scintigraphy shows that the right kidney is superior to its normal location and draining spontaneously with pelvic stasis.

Discussion

Ectopic thoracic kidney is an extremely rare congenital anomaly[1,2]. Usually, it is clinically asymptomatic and diagnosed as an incidental finding on a routine chest radiography[1]. There is a male preponderance and the majority of intrathoracic kidneys are located on the left side[3-5]. The location of thoracic ectopic kidney may be supra-, trans-, or infra-diaphragmatic[6]. The normal kidney lies beside T_{12}-L_3, its superior border ranges between T_{10} and L_1. Upper borders of thoracic kidneys range between T_7-T_{11} with transdiaphragmatic, T_7-T_9 with supradiaphragmatic, and upper pole in eventration may rise to T_5, with the kidney high but beneath the diaphragm[6]. In our case, the kidney was transdiaphragmatic and the upper pole of the kidney was at the level of T_{7-8}.

No consistent associated anomaly has been described with thoracic ectopic kidney. However, one child had trisomy 18 syndrome[3] and another patient had multiple pulmonary and cardiac anomalies in addition to the thoracic kidney[7]. Angulo et al.[2] reported a case of right intrathoracic kidney associated with a complex somite malformation that comprised vertebral fusion and right intrathoracic supernumerary ribs. There was no associated anomaly in our case.

Thoracic ectopic kidney may be a congenital anomaly or secondary to herniation through a congenital or acquired diaphragmatic defect[4,6,8].

We could not demonstrate any diaphragmatic defect in our patient.

Ebryologically, it is unclear whether the kidney ascends before the diaphragmatic leaflets close normally or if delayed diaphragmatic formation enhances the exaggerated renal ascent[8]. Initially, it was thought that intrathoracic kidneys resulted from maldevelopment of the pleuroperitoneal membrane, which caused a foramen in the posterior leaf of the diaphragm through which the kidney passed. This is now thought to be unlikely since the incidence of intrathoracic kidney with a diaphragmatic hernia is less than 0.25 percent[4]. The other possible mechanisms suggested are delayed development of the diaphragma, the influence of the developing adrenal gland and liver on renal position, and intrinsic factors in the kidney[4,6]. The synchronous association of congenital intrathoracic kidney and axial skeleton defects tends to favor the hypothesis that congenital intrathoracic kidney is secondary to persistence of the nephrogenic cord[2]. None of these mechanisms can be fully excluded and each may have a role in the development of thoracic ectopic kidney.

An intrathoracic kidney, although rare, should be considered in a child with a mass at the base of the lung on a chest x-ray. Differential diagnoses include posterior mediastinal masses such as neuroblastoma, ganglioneuroma, neurofibroma, neurogenic cysts, meningoceles, periardial cysts and Bochdalek's hernias[5]. The differential diagnosis can be made by a computed tomography (CT) that demonstrates the kidney, renal vascular pedicle and ureter[3]. IVP can demonstrate renal excretory system and ureters[3]. In our case, CT and IVP demonstrated that the right thoracic kidney was normal in size and function, but DTPA scintigraphy revealed pelvic stasis. Therefore, we suggest that DMSA and DTPA scintigraphy must be performed in cases of a thoracic ectopic kidney even if CT and IVP results are normal. No treatment is necessary once the diagnosis of congenital intrathoracic kidney has been confirmed[2].

In conclusion, although it is extremely rare, thoracic ectopic kidney should be kept in mind in differential diagnosis of a mass with a well demarcated superior margin in the lower part of the thorax.

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